

Energy Expenditure in Children with Prader-Willi Syndrome while Walking

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INTRODUCTION

- Prader-Willi Syndrome (PWS) is a rare neurodevelopmental disorder originating from an abnormality in the paternal chromosome 15q11-q13 region¹.
- Most people with PWS have increased fat mass, poor muscle mass, hypotonia, deficient growth hormone secretion, and hyperphagia¹.
- Because of the hyperphagia, low lean mass and resting metabolic rate, and high fat mass, maintaining energy balance is difficult in people with PWS^{1,2,3}.
- Walking is the most commonly reported form of physical activity in people with PWS⁴.

PURPOSE

To determine the oxygen use and caloric cost of walking at different speeds in children with PWS, and compare to non-syndromal normal weight and obese children.

METHODS

- Participants were 8 children with PWS (confirmed by genetic testing), 9 normal weight children (BMI < to the 85th percentile), 10 obese children (BMI > to the 95th percentile) (see Table 1).
- Participants completed a 10-minute rest period, a familiarization period and three 5-minute walking bouts on a treadmill at speeds of 2.0, 2.5, and 3.0 mph, with a 6-minute seated rest period in between each of the speeds.
- Body composition was measured using dual-energy x-ray absorptiometry (Lunar Prodigy Advance Plus, GE Healthcare, Madison, WI, USA). Oxygen use (VO₂) and energy expenditure (EE) normalized to body mass or lean body mass (LBM) were obtained by open spirometry (Parvo Medics TrueOne 2400, Sandy, UT, USA). Participants exercised at similar laboratory conditions (Temperature=24°C, Barometric Pressure=755 mmHg, Humidity=33%).

Table 1. Participant characteristics and physiological responses at rest, presented as mean (standard deviation) by group.

	PWS (n=8)	Normal weight (n=9)	Obese (n=10)
Male/Female	3/5	3/6	5/5
Growth hormone use	7/8		
Age (y)	11.1 (0.8)	9.8 (2.0)	10.6 (1.1)
Height (cm)	147.8 (8.2)	142.9 (20.5)	151.1 (9.6)
Body mass (kg)	44.7 (11.7)‡	35.4 (11.3)‡	62.1 (14.6)
Body mass index (kg·m ⁻²)	21.10 (6.16)‡	17.18 (1.13)‡	26.76 (3.56)
Body fat (%)	37.2 (11.4)*	22.2 (8.6)	44.5 (3.7)*
Lean body mass (kg)	26.4 (4.3)	25.5 (11.5)	33.3 (5.1)
Resting heart rate (bpm)	84 (9)	83 (13)	81 (10)
Resting VO ₂			
L·min ⁻¹	0.23 (0.05)	0.21 (0.05)‡	0.28 (0.05)
ml·kg ⁻¹ ·min ⁻¹	5.23 (0.94)	6.41 (1.26)‡	4.53 (0.61)
ml·kgLBM ⁻¹ ·min ⁻¹	8.66 (1.58)	8.81 (2.03)	8.43 (1.11)
Resting EE			
kcal·min ⁻¹	1.11 (0.25)	1.04 (0.25)	1.32 (0.25)
kcal·kg ⁻¹ ·min ⁻¹	0.03 (0.00)	0.03 (0.01)‡	0.02 (0.00)
kcal·kgLBM ⁻¹ ·min ⁻¹	0.04 (0.01)	0.04 (0.01)	0.04 (0.01)

Notes: * = different than normal weight, ‡ = different than obese; p<0.05.

RESULTS

Independent of speed PWS exhibited significantly higher heart rate than the normal weight and obese children. Children with PWS had a higher VO₂ (ml·kg⁻¹·min⁻¹) than obese (p=0.04), but similar to normal weight (p=1.00) (Figure 1). Children with PWS and normal weight children exhibited a greater caloric expenditure (kcal·kg⁻¹·min⁻¹) than obese (p=0.02 and p=0.03, respectively) (Figure 2). No differences were found among children with PWS and comparison groups for VO₂ in ml·kgLBM⁻¹·min⁻¹ or caloric expenditure in kcal·kgLBM⁻¹·min⁻¹ (Figures 3 and 4).

Figure 1 – Mean VO₂ (ml·kg⁻¹·min⁻¹) in response to walking at 2.0, 2.5, and 3.0 mph in children with PWS, normal weight children, and obese children.

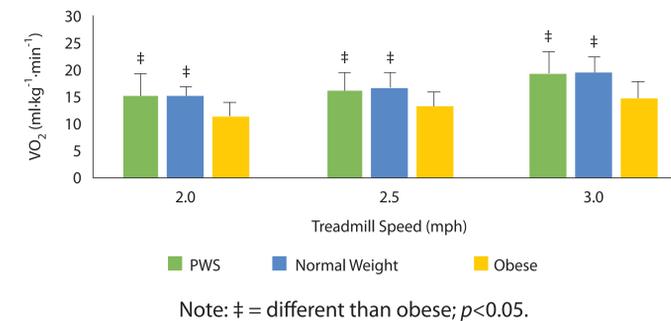


Figure 2 – Mean EE (kcal·kg⁻¹·min⁻¹) in response to walking at 2.0, 2.5, and 3.0 mph in children with PWS, normal weight children, and obese children.

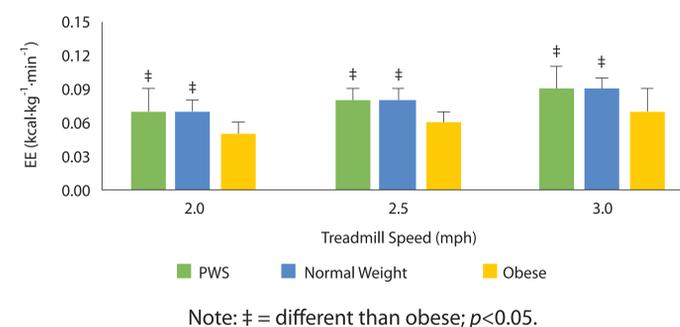


Figure 3 – Mean VO₂ (ml·kgLBM⁻¹·min⁻¹) in response to walking at 2.0, 2.5, and 3.0 mph in children with PWS, normal weight children, and obese children.

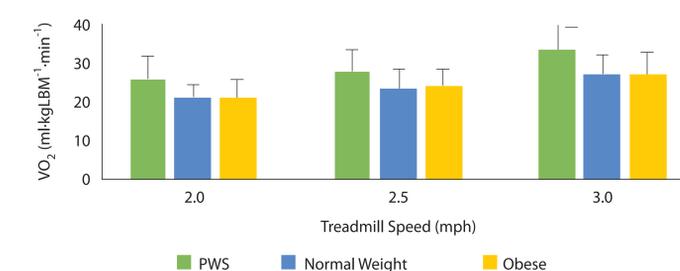
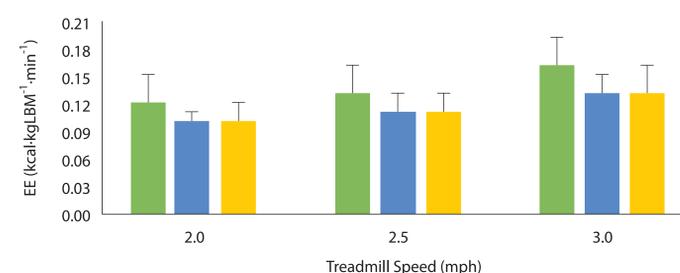


Figure 4 – Mean EE (kcal·kgLBM⁻¹·min⁻¹) in response to walking at 2.0, 2.5, and 3.0 mph in children with PWS, normal weight children, and obese children.



DISCUSSION

- Children with PWS had similar VO₂ and EE per unit of body mass as the normal weight group because of similarities in body mass. Most of these children with PWS were on growth hormone replacement therapy which helps with lean mass accrual and body fat regulation⁵, and had caloric restriction⁶.
- Obese children had a lower VO₂ and EE per unit of body mass than other groups because of their higher body mass.
- The lack of differences among the groups in VO₂ and EE when normalized for lean mass suggests comparable metabolic activity in PWS during the walking task.

CONCLUSION

- Parents of children with PWS who exhibit a normal body mass index should consider that activity energy expenditure may be the same in their children as in children without the syndrome.
- On average, a child with PWS spends 3.2 to 4.2 kcal·min⁻¹ while walking at 2.0 to 3.0 mph (at a pace of 20-30 minutes per mile). Thus, to spend 100 calories, a child this age and size would need to walk for a minimum of 24 to 31 minutes on a flat surface.

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